

Biliary Atresia

National Digestive Diseases Information Clearinghouse



National
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What is biliary atresia?

Biliary atresia is a serious but rare disease of the liver that affects newborn infants. It occurs in about one in 10,000 children and is more common in girls than in boys and in Asian and African-American newborns than in Caucasian newborns. The cause of biliary atresia is not known, and treatments are only partially successful. Biliary atresia is the most common reason for liver transplantation in children in the United States and most of the Western world.

The liver damage incurred from biliary atresia is caused by injury and loss (atresia) of the bile ducts that are responsible for draining bile from the liver. Bile is made by the liver and passes through the bile ducts and into the intestines where it helps digest food, fats, and cholesterol. The loss of bile ducts causes bile to remain in the liver. When bile builds up it can damage the liver, causing scarring and loss of liver tissue. Eventually the liver will not be able to work properly and cirrhosis will occur. Once the liver fails, a liver transplant becomes necessary. Biliary atresia can lead to liver failure and the need for liver transplant within the first 1 to 2 years of life.

What are the symptoms of biliary atresia?

The first sign of biliary atresia is jaundice, which causes a yellow color to the skin and to the whites of the eyes. Jaundice is caused by the liver not removing bilirubin, a yellow pigment from the blood.

Ordinarily, bilirubin is taken up by the liver and released into the bile. However, blockage of the bile ducts causes bilirubin and other elements of bile to build up in the blood.

Jaundice may be difficult for parents and even doctors to detect. Many healthy newborns have mild jaundice during the first 1 to 2 weeks of life due to immaturity of the liver. This normal type of jaundice disappears by the second or third week of life, whereas the jaundice of biliary atresia deepens. Newborns with jaundice after 2 weeks of life should be taken to the doctor to check for a possible liver problem.

Other signs of jaundice are a darkening of the urine and a lightening in the color of bowel movements. The urine darkens from the high levels of bilirubin in the blood spilling over into the urine, while stool lightens from a lack of bilirubin reaching the intestines. Pale, grey, or white bowel movements after 2 weeks of age are probably the most reliable sign of a liver problem and should prompt a visit to the doctor.

What causes biliary atresia?

The cause of biliary atresia is not known. The two types of biliary atresia appear to be a “fetal” form, which arises during fetal life and is present at the time of birth, and a “perinatal” form, which is more typical and does not become evident until the second to fourth week of life. Some children, particularly those with the fetal form of biliary atresia, often have other birth defects in the heart, spleen, or intestines.



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An important fact is that biliary atresia is not an inherited disease. Cases of biliary atresia do not run in families; identical twins have been born with only one child having the disease. Biliary atresia is most likely caused by an event occurring during fetal life or around the time of birth. Possibilities for the “triggering” event may include one or a combination of the following factors:

- infection with a virus or bacterium
- a problem with the immune system
- an abnormal bile component
- an error in development of the liver and bile ducts

Research on the cause of biliary atresia is of great importance. Progress in the management and prevention of biliary atresia can only come from a better understanding of its cause or causes.

How is it diagnosed?

Worsening jaundice during the first month of life means a liver problem is present. The specific diagnosis of biliary atresia requires blood and x-ray tests, and sometimes a liver biopsy. If biliary atresia is suspected, the newborn is usually referred to a specialist such as

- a pediatric gastroenterologist who is an expert in digestive diseases of children
- a pediatric hepatologist who is an expert in liver disease of children
- a pediatric surgeon who specializes in surgery of the liver and bile ducts

Initial tests. The doctor will press on the baby’s abdomen to check for an enlarged liver or spleen and order blood, urine, and stool tests to check for liver problems. The level of bilirubin in the blood will be measured and special tests for other causes of liver problems will be done.

Ultrasound of the abdomen and liver.

Ultrasound tests produce an image on a computer screen using sound waves. Ultrasound tests can show whether the liver or bile ducts are enlarged and whether tumors or cysts are blocking the flow of bile. An ultrasound cannot be used to make a diagnosis of biliary atresia, but it does help rule out other common causes of jaundice.

Liver scans. Liver scans are special types of x rays that use substances that can be detected by cameras to create an image of the liver and bile ducts. One such test is called hepatobiliary iminodiacetic acid (HIDA) scanning. HIDA scans trace the path of bile in the body and can show whether bile flow is blocked.

Liver biopsy. If another medical problem is not found to be the cause of jaundice, a liver biopsy may be recommended. For a liver biopsy, the infant is sedated and a needle is passed through the skin and then quickly in and out of the liver. A small piece of liver, about the size of a pencil lead, is obtained for examination using a microscope. A liver biopsy will usually show whether biliary atresia is likely. A biopsy can also help rule out other liver problems, such as hepatitis.

How is it treated?

Surgery. If biliary atresia appears to be the cause of the jaundice in the newborn, the next step is surgery. At the time of surgery the bile ducts can be examined and the diagnosis confirmed. For this procedure, the infant is sedated. While the infant is asleep, the surgeon makes an incision in the abdomen to directly examine the liver and bile ducts. If the surgeon confirms that biliary atresia is the problem, a Kasai procedure will usually be performed on the spot.

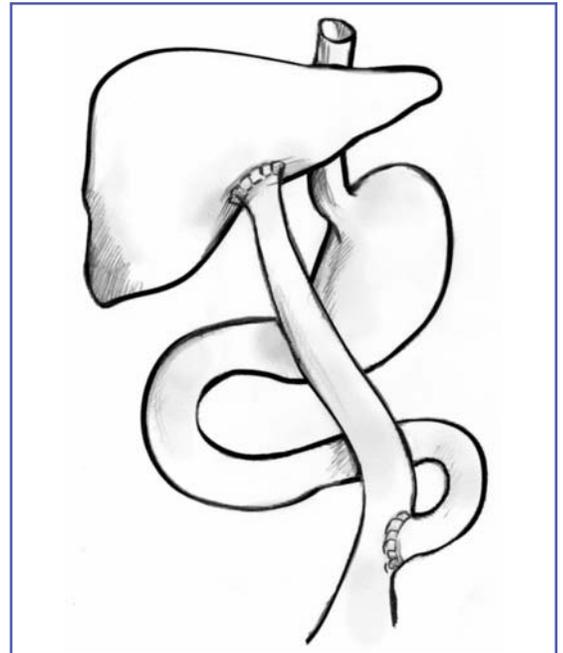
Kasai procedure (hepato-porto-enterostomy). If biliary atresia is the diagnosis, the surgeon generally goes ahead and performs an operation called the “Kasai procedure,” named after the Japanese surgeon who developed this operation. In the Kasai procedure, the bile ducts are removed and a loop of intestine is brought up to replace the bile ducts and drain the liver. As a result, bile flows from the small bile ducts straight into the intestine, bypassing the need for the larger bile ducts completely. (More about the Kasai procedure follows.)

Liver transplant. If the Kasai procedure is not successful, the infant usually will need a liver transplant within the first 1 to 2 years of life. Children with the fetal form of biliary atresia are more likely to need liver transplants—and usually sooner—than infants with the typical perinatal form. The pattern of the bile ducts affected and the extent of damage can also influence how soon a child will need a liver transplant. (More about liver transplantation follows.)

The Kasai Procedure

The Kasai procedure can restore bile flow and correct many of the problems of biliary atresia. This operation is usually not a cure for the condition, although it can have an excellent outcome. Without this surgery, a child with biliary atresia is unlikely to live beyond the age of 2. The operation works best if done before the infant is 90 days old and results are usually better in younger children.

The improved results of the surgery make the early diagnosis of biliary atresia very important, preferably before the infant is several months old and has suffered permanent liver damage. Some infants with biliary atresia who undergo a successful Kasai operation are restored to good health and can lead a normal life without jaundice or major liver problems.



The Kasai Procedure

Unfortunately, the Kasai procedure is not always successful. If bile flow is not restored, the child will likely develop worsening liver disease and cirrhosis and require liver transplantation within the first 1 to 2 years of life. In addition, the Kasai operation, even when initially successful, may not totally restore normal liver development and function. A child with biliary atresia may slowly develop cirrhosis and related complications and require a liver transplant later in childhood.

While the Kasai procedure has been a great advance in the management of biliary atresia, improvements in the operation and clinical management of children who undergo it are needed to improve the outcomes of children with this disease.

Liver Transplantation

Liver transplantation is a highly successful treatment for biliary atresia and the survival rate after surgery has increased dramatically in recent years. Children with biliary atresia are now living into adulthood, some even having children of their own. Because biliary atresia is not an inherited disease, the children of survivors of biliary atresia do not have an increased risk of having it themselves.

Improvements in transplant surgery have also led to a greater availability of livers for transplantation in children with biliary atresia. In the past, only livers from small children could be used for a child with biliary atresia because the size of the liver had to match. Recently, advanced methods have been developed to use part of an adult's liver, called "reduced size" or "split liver" transplants, for transplant in a child with biliary atresia.

In addition, surgery has been developed that allows taking part of a living adult

donor's liver to use for transplantation. Thus, parents or relatives of children with biliary atresia can donate a part of their liver for transplantation. Because healthy liver tissue grows quickly, if a child receives part of a liver from a living donor, both the donor and the child can grow complete livers over time.

Use of reduced size livers from deceased donors and left lobe livers, which are the smaller part of the liver, from living donors have greatly increased the availability of transplantation for children with liver disease. At present, almost all children with biliary atresia requiring a liver transplant will be able to receive "the gift of life," in the form of a liver from a deceased or living donor.

What happens after surgery?

Both before and after the Kasai procedure, infants will receive a specific diet with the right mix of nutrients and vitamins in a form that does not require bile to be absorbed. Poor nutrition can lead to problems with development, so doctors will monitor an infant's nutritional intake closely.

Some infants develop fluid in the abdomen after the Kasai procedure, which makes the baby's belly swell. This condition is called ascites and usually only lasts for a few weeks. If ascites lasts for more than 6 weeks, cirrhosis is likely present and the infant will probably require a liver transplant.

Also common after the Kasai procedure is infection in the remaining bile ducts inside the liver, called cholangitis. Doctors may prescribe antibiotics to prevent cholangitis or prescribe them once the infection occurs.

Children with biliary atresia may continue to have liver problems after the Kasai procedure. Even with success of the operation and return of bile flow, some children will develop injury and loss of the small bile ducts inside the liver, which can cause scarring and cirrhosis.

The liver affected by cirrhosis does not work well and is more rigid and stiff than a normal liver. As a result, the blood flow through the liver is slowed and under higher pressure. This condition is called portal hypertension. Portal hypertension can also cause flow of blood around, rather than through, the liver. This complication can cause intestinal bleeding that may require surgery and may eventually lead to a recommendation for liver transplantation.

Cirrhosis of the liver can also lead to problems with nutrition, bruising and bleeding, and itching skin. Itching, called pruritus, is caused by the build up of bile in the blood and irritation of nerve endings in the skin. Doctors may prescribe medications for itching including resins that bind bile in the intestines or antihistamines that decrease the skin's sensation of itching.

After liver transplantation, an important regimen of medicines is used to prevent the immune system from rejecting the new liver. Doctors may also continue to prescribe special diets, vitamins, blood pressure medications, and antibiotics.

Hope Through Research

Researchers are studying the possible causes of biliary atresia and new ways to diagnose and treat it. One of the largest research initiatives is the Biliary Atresia Research Consortium (BARC), a network of centers funded by the National Institute of Diabetes and Digestive and Kidney Diseases.

The network comprises 10 liver disease and transplant centers and one data-coordinating center. The centers work together to coordinate research and share ideas and resources. The network will enroll infants with biliary atresia in a large study to evaluate the best ways of managing the disease and to carry out clinical trials of new and promising treatments or approaches for diagnosis and monitoring the disease. Because biliary atresia is a rare disease, only a network of centers can identify enough infants with this disease to carry out studies of new therapies.

Centers will collect blood, tissue, and other samples from infants with biliary atresia so researchers can learn more about biliary atresia and find better treatments. An important goal of BARC is to help find the causes of biliary atresia and recommend ways for its early detection and proper management.

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The National Digestive Diseases Information Clearinghouse (NDDIC) is a service of the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK). The NIDDK is part of the National Institutes of Health under the U.S. Department of Health and Human Services. Established in 1980, the Clearinghouse provides information about digestive diseases to people with digestive disorders and to their families, health care professionals, and the public. The NDDIC answers inquiries, develops and distributes publications, and works closely with professional and patient organizations and Government agencies to coordinate resources about digestive diseases.

Publications produced by the clearinghouse are carefully reviewed by both NIDDK scientists and outside experts. This fact sheet was reviewed by BARC investigators: Ronald Sokol, M.D., University of Colorado/The Children's Hospital of Denver; Jorge Bezerra, M.D., Cincinnati Children's Hospital and Medical Center; and Benjamin Shneider, M.D., Mount Sinai Hospital of New York. The original illustration of the Kasai procedure was provided by Julie Porter.

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